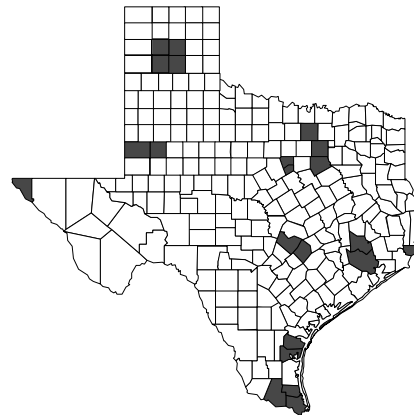


**BIRTH DEFECT INVESTIGATIONS CONDUCTED BY  
THE TEXAS BIRTH DEFECTS MONITORING DIVISION AND/OR THE HEALTH  
STUDIES PROGRAM, TEXAS DEPARTMENT OF HEALTH**

**1996**

**Notes:** A glossary of birth defects and other terms is found at the end of this document.  
TDH = Texas Department of Health.  
Because Texas does not yet have a state-wide Birth Defects Registry, expected rates were calculated using data from the California Birth Defects Monitoring Program.

Counties In Which Investigations Were Conducted



**HOOD, SOMERVELL AND ELLIS COUNTIES**

**Condition of Concern:** *Down syndrome.*

**Background:** TDH received a call from the Texas Department of Mental Health and Mental Retardation of an apparent excess number of Down syndrome cases from Granbury (Hood County). Seven cases were observed, born from 1992 - 1994. Based on the California rate, this was 14.3 times the number that was expected in the county, and was statistically significant (i.e. the excess had a low probability of having arisen by random variation). Four months later, TDH received a call from a concerned citizen about eight cases of Down syndrome in Ellis County, born from 1992 to 1994. This was 1.9 times times the expected number and was not statistically significant.

**Response:** Because the clusters were geographically close to each other and occurred at the same time, the investigations were combined. The TDH investigations attempted to (a) confirm reported cases and (b) ascertain and confirm any unreported cases for the areas. Five cases were confirmed, born to mothers resident in Hood County. This was 4.3 times the expected number, and was statistically significant. In Somervell

County, one confirmed case was 4.9 times the expected number, but was not statistically significant. There were 12 confirmed cases in Ellis County, which were 2.8 times the number expected and statistically significant. For all three counties, the 18 cases were 3.2 times the expected number. Because the total number of confirmed cases showed a statistically significant excess, case mothers were interviewed to determine if there were any risk factors in common that might explain the clusters. The excess observed for each county was not explained by maternal age, race/ethnic distribution, or other known risk factors for Down syndrome. The investigation did not provide evidence that environmental factors were associated with the excess occurrence of Down syndrome cases, but its ability to do so was limited.

**Duration of Investigation:** September 1994 - May 1996. 388 person-hours in 1996; 1,280 person-hours total.

**GAINES AND DAWSON COUNTIES**

**Condition of Concern:** *Cleft lip and palate.*

**Background:** In March of 1995, TDH was contacted by a citizen concerned about a potential excess of cleft lip and palate in Gaines County. Three cases were reported. Based on California's rates, this was 16.0 times the number of cases expected and statistically significant. Suspicion of excess cases in neighboring Dawson County resulted in expanding the investigation to include it as well.

**Response:** TDH is conducting an investigation to (a) confirm reported cases and (b) ascertain and confirm any unreported cases for the county. The investigation is ongoing.

**Duration of Investigation:** March 1995 - present. 21 person-hours in 1996; 36 person-hours total.

#### **DAWSON COUNTY**

**Condition of Concern:** *Spina bifida.*

**Background:** During the investigation of cleft lip and palate in Gaines County, Early Childhood Intervention staff mentioned concern of high numbers of spina bifida cases in the next county to the east, Dawson. Three cases were reported to have been born in 1994. Based on California's rates, this was 24.4 times the number of cases expected and statistically significant.

**Response:** TDH attempted to (a) confirm the reported cases and (b) ascertain and confirm any unreported cases for the county. TDH was not able to confirm any of the cases. No further action required at this time.

**Duration of Investigation:** March 1995 - March 1996. 2 person-hours in 1996; 12 person-hours total.

#### **THE WOODLANDS (Montgomery Co.)**

**Condition of Concern:** *Anencephaly.*

**Background:** Concerned citizens collected information on potential anencephaly cases in and around The Woodlands for several years. They offered this information to TDH, to assist with investigating a possible cluster in the area.

**Response:** Using the information, TDH conducted an investigation to (a) confirm the reported cases and (b) ascertain and confirm any unreported cases for the area. In The Woodlands from 1990-1994, the three confirmed cases of anencephaly were 3.1 times what would have been expected, but this was within the limits of random variation. In the same time period in Montgomery County as a whole, 10 confirmed cases were 2.0 times the expected number. When race/ethnic group was considered, the ratio increased to 2.3 times, and became statistically significant.

**Duration of Investigation:** August 1995 - October

1996. 165 person-hours in 1996;  
217 person-hours total.

#### **HOUSTON (Harris Co.)**

**Condition of Concern:** *Down syndrome.*

**Background:** A large hospital in Houston reported a possible excess number of Down syndrome cases delivered at the facility. Six children with Down syndrome were born in June and July of 1995. If there had been the same frequency of occurrence as in the prior 10 months, we would have expected only 1.6 cases. (Because this is a hospital-based cluster, it was not compared to expected values derived from population-based registries, such as California.)

**Response:** Further case finding and case verification were deemed unnecessary in this hospital-based cluster. The six cases were 3.8 times the number of expected cases, based on routine nursery surveillance (i.e., the genetics logs). This was statistically significant. Since this cluster occurred within the pilot study area, Texas Birth Defects Monitoring Division staff will monitor Down syndrome births at the facility. No further action required at this time.

**Duration of Investigation:** August 1995 - December 1996. 2 person-hours in 1996; 40 person-hours total.

#### **CAMERON, WILLACY AND HIDALGO COUNTIES**

**Condition of Concern:** *Ventricular septal defects.*

**Background:** A health practitioner shared study results which suggested that muscular type ventricular septal defect (VSD) rates seen in a clinic were significantly higher in the Lower Rio Grande Valley when compared to other parts of the country or to other studies. It was also reported that VSD rates were higher compared to the prior year in the same clinic.

**Response:** TDH reviewed the study, and compared it with similar studies in the scientific literature. It was very difficult to establish the existence of excessive rates using data from the study, as it would be from any referral center in general. This was due to problems in assessing the base population, and to the impact of referral patterns, improved case detection using modern diagnostic tools, and case counting. No further action required at this time.

**Duration of Investigation:** January - February. 16 person-hours.

#### **VIDOR (Orange County)**

**Condition of Concern:** *Multiple birth defects.*

**Background:** A citizen called to report the 1995 birth of a child with sirenomelia and another child with polycystic kidneys. There was concern regarding the environmental health impact of a paper mill.

**Response:** The citizen had called the Bureau of Vital Statistics for data on birth defects in that area (which was not yet covered by the Birth Defects Registry). Also, referral was made to the Texas Teratogen Information Service, for information about paper mills. No further action required at this time.

**Duration of Investigation:** February - September.  
2 person-hours.

### HOOD COUNTY

**Condition of Concern:** *Chromosomal defects.*

**Background:** An employee of a local health department contacted TDH regarding an apparent excess of trisomy 13 (Patau syndrome), trisomy 18 (Edwards syndrome), and Turner syndrome. Turner syndrome is not routinely reported by comparison birth defect registries, so was dropped from the investigation. The two reported cases of trisomy 13 were 7.4 times expected, and the excess was close to achieving statistical significance. The one case of trisomy 18 was 2.2 times expected, and was not significant.

**Response:** A search of fetal death and infant death certificates was made to confirm cases and ascertain any that had been missed. Only one case was confirmed for each trisomy. Thus occurrence of trisomy 13 was 3.7 times expected, and trisomy 18, 2.2 times expected; both numbers were within the limits of random variation.

**Duration of Investigation:** March - April.  
7 person-hours.

### LEWISVILLE (Denton County)

**Condition of Concern:** *Multiple birth defects and conditions.*

**Background:** A concerned parent contacted TDH about children born in a neighborhood near a swimming pool manufacturer. The children suffered from visual or hearing impairment, mental retardation, or heart defects.

**Response:** There was a discussion regarding the approach used in investigating clusters, and the difficulty of doing so when there is a variety of conditions involved. The parent was asked to obtain more detailed information on the reported cases, but has not sent the information. No further action required at this time.

**Duration of Investigation:** April - December.  
2 person-hours.

### STATE OF OKLAHOMA

**Condition of Concern:** *Renal agenesis.*

**Background:** A staff member of the Oklahoma Birth Defects Registry called regarding 6-8 cases of renal agenesis in Oklahoma residents close to the OK/TX border. Help was requested from TDH in getting information from Texas hospitals in which Oklahoma residents may have delivered.

**Response:** TDH identified the number of livebirths to OK residents in hospitals close to the Oklahoma border. The Oklahoma registry then discovered that the initial cluster concern may have been an artifact of reporting practices and misdiagnosis. No further action required at this time.

**Duration of Investigation:** April - August.  
5 person-hours.

### PANTEX HEALTH ASSESSMENT (Armstrong, Carson, Potter and Randall Counties)

**Condition of Concern:** *All birth defects.*

**Background:** TDH was invited by the Agency for Toxic Substances and Diseases Registry (ATSDR) to conduct a health assessment in the area surrounding Pantex, a nuclear weapons facility. Citizens had voiced concerns regarding a variety of health conditions including birth defects.

**Response:** Cases of structural birth defects delivered from 1990-1994 were identified by examining birth, fetal death and death certificates. The numbers observed were compared to numbers expected, using rates derived from the entire State of Texas for each of the certificate types. It was found that there were no consistently significant elevations of birth defects among fetal death certificates or death certificates. There were several types of defects that appeared elevated when examining birth certificates. However, more careful reporting of birth defects could not be ruled out as the cause of these excess rates. There was no parental occupation that appeared to be frequent among parents of birth defect cases. There was no consistent pattern showing that closer proximity to Pantex increased the risk of birth defects.

**Duration of Investigation:** May - September.  
127 person-hours.

### FABENS (El Paso County)

**Condition of Concern:** *Heart defects.*

**Background:** A family physician contacted TDH, concerned that he was seeing an abnormally high number of children with heart defects. The time period and geographical area of concern remain to be determined.

**Response:** TDH staff will visit the physician's office to abstract medical records. This will allow more precise definition of the extent and nature of the concern. The investigation is ongoing.

**Duration of Investigation:** July - December.  
18 person-hours.

#### **KINGSVILLE (Kleberg County)**

**Condition of Concern:** *Multiple defects and health conditions.*

**Background:** A citizen contacted TDH regarding a variety of defects and health conditions in children born to women working in the same office. She was particularly concerned that the children's health problems might have been related to installation of a fire retardant in the office walls.

**Response:** Occurrence of each defect or condition was compared to what might have been expected, though this was based on births in the county. There were too few cases of any particular condition to achieve statistical significance or to allow further investigation. The timing of exposure and the variety of health outcomes made it unlikely that the fire retardant or its solvent were responsible for all the conditions reported. No further action required.

**Duration of Investigation:** August - September.  
17 person-hours.

#### **GRAND PRAIRIE (Dallas County)**

**Condition of Concern:** *Multiple defects.*

**Background:** A concerned parent contacted TDH regarding a perceived excess of miscarriages and birth defects in a subdivision of Grand Prairie, TX from 1994 to 1996. The reported birth defects included one case of Klinefelter syndrome and two cases of Down syndrome. It is difficult to make meaningful conclusions from one case. The Down syndrome cases were fewer than the 2.69 cases that would have been expected in Grand Prairie as a whole.

**Response:** No further action regarding birth defects is required at this time. TDH will respond concerning the miscarriages.

**Duration of Investigation:** August - December. 4 person-hours.

#### **NUECES COUNTY**

**Condition of Concern:** *Anencephaly*

**Background:** In October, TDH received a call from a citizen concerned about a possible link between a large petrochemical company in Corpus Christi and three cases of anencephaly. The time period of primary concern was 1990-1993.

**Response:** Using information from vital records, TDH tabulated the number of anencephaly cases in Nueces County. That was compared to the number which would have been expected from state-wide rates for 1981-1989 and 1990-1993. Those rates were also generated from vital record data. Based on state-wide rates, 24.6 cases of anencephaly would have been expected to occur in Nueces County for 1981-1989. Twenty-one cases were reported on vital records. For 1990-1993, 7.6 cases were expected with 12 actually observed. Although more cases of anencephaly were observed during the later time period than would have been expected, the observed to expected ratio (1.58) was within the limits of random variation. No further action required at this time.

**Duration of Investigation:** October - December.  
22 person-hours.

#### **TRAVIS AND BASTROP COUNTIES**

**Condition of Concern:** *Anophthalmia.*

**Background:** TDH was contacted by a staff member of Early Childhood Intervention about four cases of anophthalmia born from June-October 1996 in Travis and Bastrop Counties. Based on rates from the California Birth Defects Monitoring Division, this was 20.2 times the number expected and statistically significant.

**Response:** TDH is conducting an investigation to (a) confirm reported cases and (b) ascertain and confirm any unreported cases for the county. The investigation is ongoing.

**Duration of Investigation:** October - December.  
10 person-hours.

## GLOSSARY

|                              |   |
|------------------------------|---|
| Anencephaly:                 | congenital absence of the skull.  |
| Anophthalmia:                | abnormal development of the eye.  |
| Atresia:                     | imperforation; absence or closure of a normal opening.  |
| Chromosomal defects:         | birth defects resulting from abnormal chromosomes, including for example Down syndrome (extra chromosome #21), Patau syndrome (extra chromosome #13), or Turner syndrome (only one X chromosome instead of two Xs or an X and Y). |
| Cleft lip:                   | the congenital failure of the maxillary and median nasal processes to fuse, forming a groove or fissure in the lip; also called harelip.  |
| Cleft palate:                | the congenital failure of the palate to fuse properly, forming a grooved or fissured depression in the roof of the mouth.   |
| Down syndrome:               | the chromosomal abnormality that is characterized by moderate-to-severe mental retardation, sloping forehead, small ear canals, flat-bridged nose, and short phalanges. Due to an extra chromosome 21; one type of trisomy.       |
| Gastroschisis:               | a congenital fissure of the abdominal wall with protrusion of the intestines.   |
| Holoprosencephaly:           | impaired midline cleavage of the embryonic forebrain, the most extreme form being cyclopia.   |
| Klinefelter syndrome:        | an abnormal condition in a male characterized by one Y chromosome and two or three X chromosomes, small testicles and infertility.  |
| Neural tube defects: (NTDs)  | a general term for a number of defects which are presumed to have a common origin in failure of the neural tube to develop properly during the embryonic stage. Conditions include among others, anencephaly and spina bifida.    |
| Person-hour:                 | the equivalent of one person working for one hour; thus 3 person-hours can be derived from 3 people working 1 hour each, or 1 person working 3 hours, or any other combination.   |
| Polycystic kidneys:          | a hereditary congenital condition characterized by bilateral multiple renal cysts.  |
| Renal agenesis:              | congenital absence of one or both kidneys.  |
| Sirenomelia:                 | union of the legs with partial or complete fusion of the feet.  |
| Spina bifida:                | a developmental anomaly characterized by defective closure of the bony encasement of the spinal cord, through which the cord and meninges may or may not protrude.  |
| Trisomies:                   | having three copies of a chromosome instead of the usual two.   |
| Trisomy 13 (Patau syndrome): | the chromosomal abnormality that is characterized by impaired midline   |

|                                |   |
|--------------------------------|---|
|                                | facial development, cleft lip and palate, polydactyly, and mental retardation. Due to an extra chromosome 13; one type of trisomy.  |
| Trisomy 18 (Edwards syndrome): | the chromosomal abnormality that is characterized by mental retardation, neonatal hepatitis, low-set ears, skull malformation, and short digits.  |
| Trisomy 21:                    | see Down syndrome.  |
| Turner syndrome:               | a genetically determined condition that is associated with the presence of one X chromosome and no Y chromosome and characterized by an outwardly female appearance with incomplete and infertile gonads. |
| Ventricular septal defect:     | a congenital heart defect in which there are one or more holes in the wall dividing the two ventricles of the heart.  |

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*For copies of this report or for further information, please call the Texas Birth Defects Monitoring Division, Texas Department of Health at (512) 458-7232.*